# Hypertrophic Spinal Pachymeningitis in a Patient with Chronic Antineutrophil Cytoplasmic Antibody-Associated Nephritis: A Case Report

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Hypertrophic spinal pachymeningitis (HSP), a rare condition characterized by a thickened inflamed dura mater, is caused by antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis[1)]. Symptoms progress from local pain to myelopathy[2)]. We report a case of HSP in a patient with long-term ANCA-related nephritis treated surgically and with corticosteroids.

A 69-year-old Japanese man presented to the orthopedic clinic with progressive lower extremity numbness, weakness, and gait disturbance. He was diagnosed with ANCA-related nephritis and diabetes 10 years earlier. He had undergone steroid pulse therapy three times and was maintained on 2-8 mg of prednisolone when the nephritis worsened and the myeloperoxidase-ANCA antibodies increased (Fig. 1). Magnetic resonance imaging (MRI) 5 years earlier showed no spinal cord compression (Fig. 2A). Upon presentation, lower extremity muscle power was grades 0-2/5 (right) and 0-2/5 (left). Sensory disturbance affecting pain and touch was detected below T10. Serologic tests, including white blood cell count and C-reactive protein levels, were normal. MRI revealed a T3 dorsal mass compressing the spinal cord (Fig. 2 B, C). Contrast-enhanced computed tomography revealed mass enhancement (Fig. 2D). The diagnosis was undetermined preoperatively, and a pathologist was consulted for intraoperative rapid analysis.

Laminectomy was performed at T1-3 (Fig. 3A). Ultrasonography revealed a thickened dura mater (Fig. 3B). Intraoperative rapid pathologic analysis of the excised dura revealed inflammatory cells but no tumor cells. Because the findings were compatible with HSP, the dura mater was partially resected (Fig. 3C, D). Hematoxylin and eosin staining showed dense collagenous thickening of the dura mater with focal chronic inflammatory infiltrates (Fig. 3E, F), and Elastica-Masson staining revealed obstructive vasculitis limited to the small veins (Fig. 3G, H), supporting a diagnosis of chronic HSP. Steroid pulse therapy was administered 2 weeks after surgery. Lower extremity numbness and weakness improved immediately after surgery. Within 6 months, the patient could walk with a cane. MRI revealed that the dorsal mass remained 1 week after surgery (Fig. 4A) but disappeared after 3 months of steroid pulse therapy (Fig. 4 B) and no recurrence was observed 1 year after surgery (Fig. 4C).

Few cases of HSP-related myelopathy symptoms with chronic ANCA-associated nephritis have been treated with combined surgical and corticosteroid therapies. This report highlights the development of HSP even with long-term steroid administration and three pulse therapies. If pathologic analysis reveals inflammation and vasculitis, HSP may improve with steroid pulse therapy.

HSP often presents with acute clinical symptoms of ANCA-associated nephritis[3-5)]. In such cases, corticosteroids are the mainstay of nonoperative treatment[4)]. Corticosteroids may control the inflammatory process and improve dural thickening with prolonged therapy. Ogaki et al. reported a patient with acute-onset myeloperoxidase-ANCA-related HSP who experienced short-term recurrence after receiving only steroid treatment[5)]. In some cases of ANCA-related

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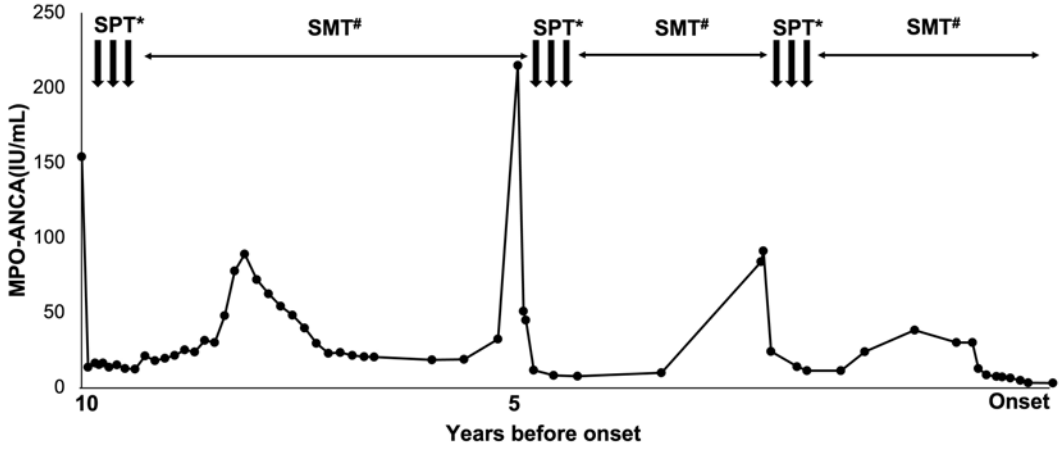


**Figure 1.** Time course of myeloperoxidase-anti-neutrophil cytoplasmic antibody (MPO-ANCA) and steroid therapy. *SPT indicates steroid pulse therapy with prednisolone 20 mg/day. #SMT indicates steroid maintenance therapy with prednisolone 2–8 mg/day.

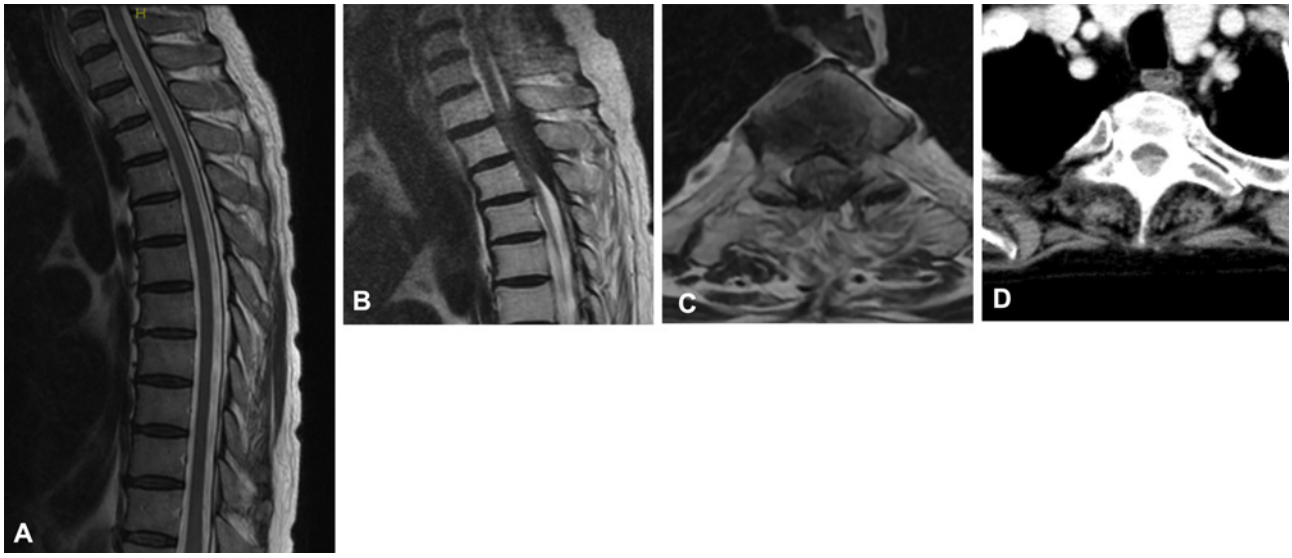


**Figure 2.** MRI and contrast-enhanced computed tomography (CT) images. Sagittal images 5 years before onset (A). Sagittal and axial images at onset (B and C). Contrast-enhanced CT images at onset (D).

HSP treated with steroids and immunosuppressive therapy, control is not achieved[6]. Therefore, decompression surgery with excision of the thickened dura is the treatment of choice in most cases with neurologic symptoms[4].

The pathologic findings of inflammation and vasculitis of the dura mater suggest the potential effectiveness of steroid treatment in acute HSP[7,8]. Nagashima reported a case of vasculitis of unknown etiology; inflammation was limited to the veins, with the arteries remaining unaffected[7]. Pathologic examination in this case with a long-term course of ANCA-associated nephritis revealed inflammation, in contrast to previous acute-onset cases[3-5]. The patient was highly responsive to steroids, and combination therapy was administered without total dura mater resection. In cases of suspected HSP owing to chronic ANCA-related nephritis, intraoperative pathology should guide steroid therapy planning. A review of HSP suggests that active inflammation of the dura preoperatively may contribute to recurrence[9]. Our case requires careful follow-up.

**Conflicts of Interest:** The authors declare that there are no relevant conflicts of interest.

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**Author Contributions:** GK designed the study. KW, YN, KA, and AK corrected the manuscript. YI supervised the study. GK wrote the manuscript.

**Ethical Approval:** This study was approved by the Ethics Committee of the Hirosaki University Graduate School of Medicine (approval code: 2022-072).

**Informed Consent:** Informed consent for publication was obtained from the patient described in this study.

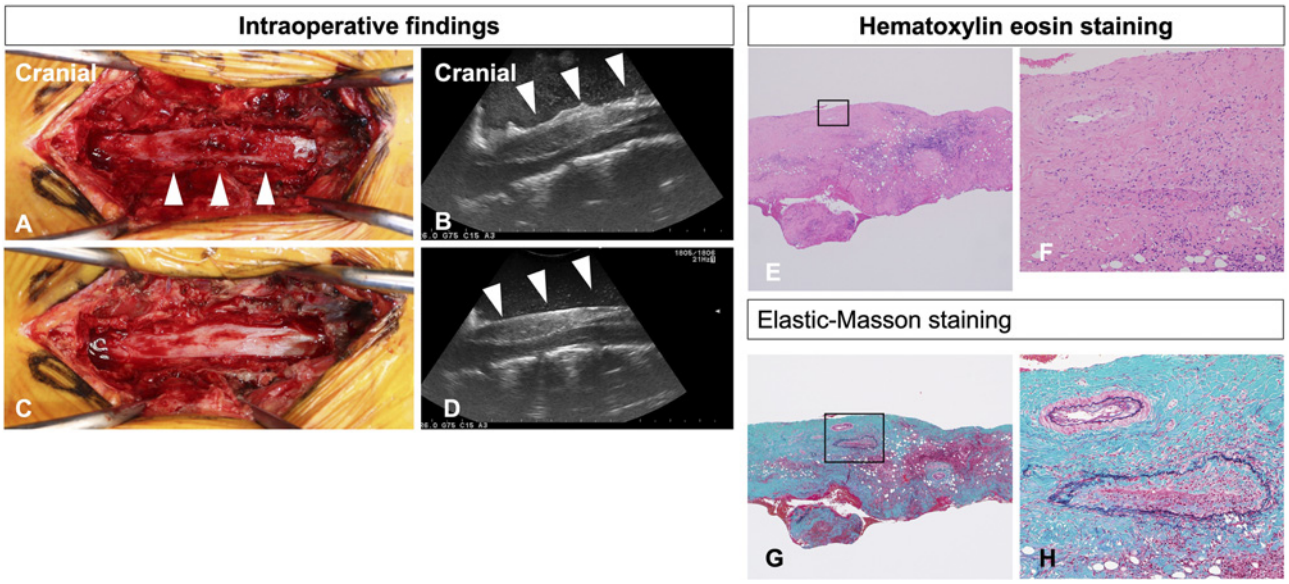


**Figure 3.** Laminectomy performed at the T1-3 level (A). Ultrasonography before dura resection (B). Dura mater partially resected (C). Ultrasonography after dura resection (D). Arrowhead indicates thickened dura mater. Hematoxylin eosin staining shows thickened dura matter and focal chronic inflammatory infiltrates (E and F), and Elastica–Masson staining showed evidence of phlebitis obliterans (G and H) with intact arteries close to the affected vein. F and H are the high-magnification images of the black boxes in E and G, respectively.

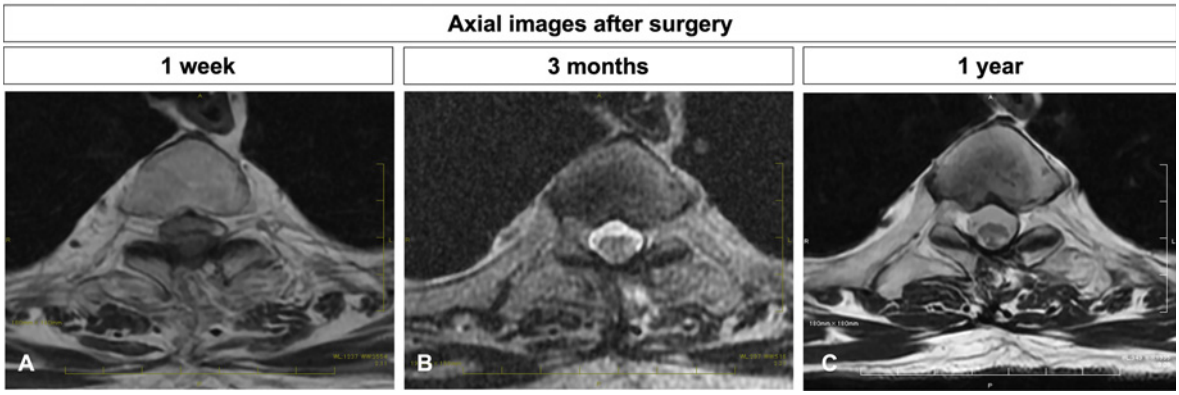


**Figure 4.** MRI axial image showing that the dorsal mass remained a week after surgery (A) and disappeared 3 months after steroid pulse therapy (B). MRI axial image shows no recurrence 1 year after surgery (C).

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